Chisolm (J. J.) With Compliments of J. J. C.

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Intra-Ocular Enchondroma

OF

TWENTY-TWO YEARS' GROWTH,

Removed at the Baltimore Eye and Ear Infirmary.

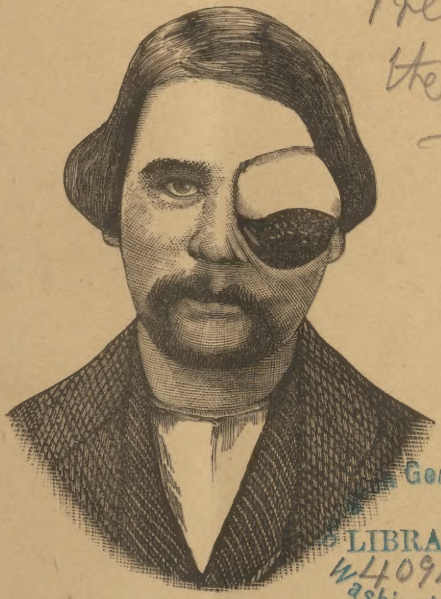
By J. J. CHISOLM, M.D.,

Clinical Professor of Eye and Ear Surgery in the University of Maryland.

WITH A DESCRIPTION OF THE MICROSCOPIC CONDITION OF THE TUMOR,

By H. KNAPP. M.D.,

Presented by the Author



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WITH A DESCRIPTION OF THE MICROSCOPIC CONDITION OF THE TUMOR.

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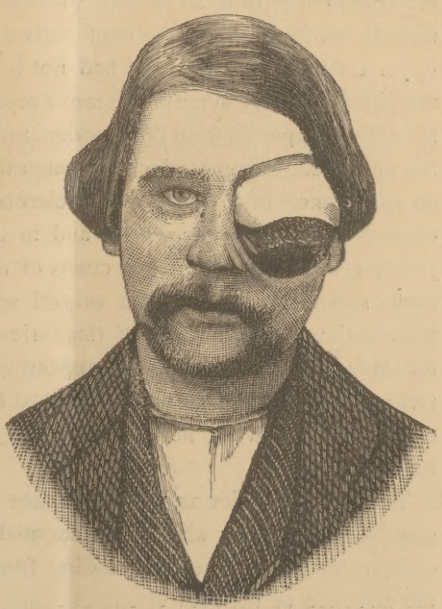
OF NEW YORK.

S. H. V———, of Virginia, aged twenty-five, was born a healthy infant. At the age of three years his parents noticed a difference in the size of his eyes, the left one being slightly enlarged. Upon further examination and by the use of tests, the sight in it was found defective. Prior to this discovery both eyes were supposed to be perfect. From that period until the present, covering an interval of twenty-two years, the left eyeball has continued steadily to increase in size, at times accompanied by pain. He has been pursuing the avocation of a farmer, and continued work until he left home to seek surgical advice. In his desire to get rid of this projecting and horrible deformity he entered the Baltimore Eye and Ear Infirmary for surgical treatment.



His general health is good, height 5 ft. 10 in., weight about 130 lbs. The right eye is perfect. In the place of the left eye, a rounded mass protrudes (see figure, from a photograph), which seems to be as large as the fist. After appar-

ently filling the orbital cavity, the growing tumor has gradually projected forwards until it has attained a prominence of 2½ inches from a level of the face, having about the same dimensions of 2½ inches in its vertical and transverse diameter. An immensely expanded, hypertrophied, and movable upper lid, as a broad curved belt, covered the upper three-fourths of the free surface of the projection. In this overgrown lid the orbicular muscle was well developed, as indicated by the numerous crescentic ridges which encircled the facial base of the tumor, and which were made prominent through the subcutaneous bundles of muscular fibres. On the upper and outer surface of the tumor, apparently under the thickness of the lid substance, was an elastic, soft swelling, supposed to be the lachrymal gland, pushed out of its natural site, and drawn forward by the growing mass. The mucous or conjunctival tissue covering the exposed face of the tumor was thickened, but was otherwise apparently healthy, possessing the usual mobility of this membrane over the sclerotic coat of the eye. The lower conjunctival cul-de-sac had been effaced in the continued requirements of the enlarging growth, so that the Meibomian edge of the lower lid was directly continuous with the conjunctival covering of the tumor. The upper conjunctival cul-de-sac had not been altogether obliterated, but remained as a very superficial groove separating the edge and inner surface of the upper lid from the anterior surface of the protrusion. Under the ocular conjunctiva, which, although thickened, was still translucent, an appearance of healthy, white sclerotic was visible. All traces of corneal tissue had disappeared, and in its stead was a thick, red conjunctiva, covering which were crusts of desiccated mucus. From this fleshy surface he reports that several serious hemorrhages have from time to time taken place. Of this serious complication I could detect no evidences, as there was no appearance of ulceration when the anterior surface of the tumor was exposed by the removal of the mucous crusts. There were no large vessels visible on the surface of the tumor.

The entire anterior and exposed face of the growth was kept constantly bathed by an abundant lachrymal secretion, which, in a certain oblique light, could be seen oozing from beneath the upper lid, from the direction of the soft, elastic swelling already noticed as lying upon

the upper and outer surface of the tumor, and supposed to be the lachrymal gland.

The long-continued pressure of this ever-slowly increasing growth had caused, by absorption, an increase in size and a change in direction of the outer border of the orbit. It had also in a measure displaced the nose from its median position. After leaving the orbit the growth had not spread out suddenly, becoming bulbous, but had retained a uniform diameter in every direction of about two inches and a half, which measured the degree of expansion of the orbital opening.

The whole tumor, which was uniformly ovoidal in shape, *possessed limited movements, synchronous with those of the sound eye.* When firmly held and a to-and-fro movement imparted to the hand, similar movements were extended to the growth. Careful palpation elicited no fluctuation, although a decided feeling of elasticity pervaded the entire tumor.

The *history, symptoms, and appearance of the projecting mass indicated an intra-ocular tumor* of very slow growth, which, after twenty-two years of steady increase, was still incarcerated within the chambers of the eye, and isolated by the much expanded and thickened, but, to all appearances, otherwise normal sclerotic coat. Believing that the eye was the seat of a malignant growth, which, from some unexplainable cause, had taken on a very irregular and unusual development, its extirpation was advised and assented to. The operation, which was performed under chloroform, in the presence of Prof. Gross, of Philadelphia, and others, was similar to the ordinary enucleation so familiar to ophthalmic surgeons. After extending the outer canthus, on account of the great size of the tumor, the conjunctiva was incised around the former site of the cornea, the tenotomy of the various muscles was easily effected, and after passing a long, heavy scissors, curved on the flat near the point, to the very apex of the conical cavity of the orbit, the optic nerve was divided, and the whole mass at once glided out of the socket, leaving a huge but apparently healthy cavity, devoid of all fat and lined from rim to apex with a thickened periosteum. Hemorrhage, not excessive, was readily controlled by sponge pressure.

The case made good progress for the first few days. On the fourth

night from the operation there was a hemorrhage of a few ounces. On the fifth night this was again repeated without the source of hemorrhage being discovered. On the evening of the ninth day a very severe hemorrhage took place, which was deemed sufficiently serious to warrant me in ligating the common carotid. On the morning of the tenth irregular tetanic symptoms supervened, and he died on the fourth day after the ligature had been applied. There had been no recurrence of the hemorrhage.

The mass removed was a smooth, regularly ovoidal tumor, about three and a half inches in its longest diameter, corresponding with the axis of the orbit, and two inches and a half in its vertical and transverse diameters. Its outer surface was the expanded and thickened sclerotic coat of the eye, which apparently had not been broken through at any point by the growth from within. To this outer surface were attached the muscles of the eyeball. When the tumor was laid open the whole sclerotic sac was found filled with a solid contents, mottled in appearance, and of varied consistence, among which were conspicuous white nodules of different sizes. These proved upon section to be cartilage, which character was confirmed by microscopic examination.

It is so very unusual to find cartilage in the growths within the eyeball, that this rare specimen of tumor was deemed worthy of a more careful examination. Prof. H. Knapp, a recognized authority on intra-ocular growths, has very kindly consented to make the examination for me, and to him are we indebted for the following carefully prepared and most valuable report of this rare pathological specimen:—

ANATOMICAL EXAMINATION OF THE PSEUDOPLASM.

The tumor (Fig. 1, Tab. A), hardened in Mueller's fluid, was roundish, having a diameter of two inches and a half. It was surrounded by connective tissue in the form of an irregularly dense *capsule* (Fig. 1, *a*). On the transverse section its bulk consisted of *hard nodes* (Fig. 1, *b*), separated from one another by *fibrous tissue* connecting with the capsule. A considerable portion of the tumor, about one-fifth of its size, was softer and had a *fibro-granular* appearance (*c*).

The enveloping *capsule* consisted in its outer part of dense undulating and parallelly striated connective tissue, containing a very scanty amount of cells and blood-vessels. Its similarity with the structure of the sclerotic was marked. The inner layers of the capsule had more blood-vessels and cells, and the layer near the nodes was filled with small round formative cells, scattered sparsely through the fibres; besides them smaller and larger cells lay together in spindle-shaped spaces between the bundles of fibres. These layers, therefore, presented the connective tissue in the state of growth or proliferation.

The *fibro-granular* part (*c*) of the tumor showed a net of connective-tissue fibres (Fig. 2) which was freely traversed by blood-vessels (*v*), and abundantly interspersed with formative cells (*f*). In addition to these elementary parts this portion of the tumor contained a good deal of fat, in the form of globules (*m*), and granulated bodies (*g*). Teased preparations of this substance showed long fibres with double outlines (*o*), spindle-shaped (*s*), stellate (*h*), and small round

cells (*r*), the latter here and there in dense clusters (*u*).

The structure of the hard *nodes* discovered the two varieties of *cartilage*, the *hyaline* and the *fibrous*, in very characteristical pictures. The *hyaline* variety (Figs. 3, 4) prevailed over the fibrous (Fig. 5). It consisted of a yellowish, homogeneous, or very finely granular, glassy-looking matrix—intercellular substance—in which cartilage-cells of different forms were embedded. Oval cells with one or two nuclei (Fig. 3, *a*); small, round, oval and irregular spaces containing three and more nuclei (*b*); large, irregular cavities densely filled with nuclei (*c*), and free nuclei (*d*), were promiscuously placed in the homogeneous hyaline basis-substance. In many places the matrix appeared interspersed with free nuclei alone (Fig. 4). Part of the nuclei were small, round and pellucid (*d'*); another part were small and shrivelled up (*b*); others (*cd*) large, oval, round, club-like, dumb-bell shaped or irregular. Their interior was either homogeneous and transparent (*c*), or finely or coarsely dotted (*d*).

The *fibrous portion of the cartilage* (Fig. 5) showed an irregularly striated matrix in which the same cellular elements were embedded as in the hyaline portion. Where the nuclei predominated (*a*), the matrix had commonly a granular appearance, with fine, short lines; where the larger cells were prevalent (*b*), the matrix was markedly fibrous.

Blood-vessels ran in different directions through the fibrous part, but were absent in the hyaline portion.

From the foregoing description it is evident that the

tumor was, as Dr. Chisolm declared it to be, an *enchondroma.* There is, further, no doubt that it was, from beginning to end, an *intra-ocular* growth. The history showed the gradual enlargement of the eyeball, together with failure of sight. Had the enlargement been only apparent, due to exophthalmus, in consequence of an orbital tumor, no impairment of sight would have been noticed at the outset of the difficulty, for we see the protrusions of the eye reach very high degrees before the vision becomes impaired. The fact that the sclerotic coat, before and after the operation, was seen to envelop the growth; furthermore, the movements of the tumor concomitant with those of the other eye; the insertions of the ocular muscles into the surrounding coat—sclerotic; their division in the enucleation of the eye; the gliding of the whole mass out of the orbit after the muscles and the optic nerve had been severed:—all these circumstances show *that the growth was within the eye.* As far as I can ascertain, this case is the only one of its kind on record. I have consulted many older and modern works on morbid growths, as well as the text-books of ophthalmology, without finding a mention of intra-ocular enchondroma.

As this case introduces a new species of tumor into the field of ophthalmology, I have endeavored not only to *define its character*, but to *trace its origin and development.* If we look for analogy, and remember that cartilaginous tumors, though originating mostly in the bone and periosteum, not infrequently arise from the connective tissue of other organs, especially the fasciæ, (for instance in the parotid gland), we may assume that

the tissue in which, most likely, the above-described enchondroma took its first root, was the *sclerotic.* The exceedingly slow increase of the tumor also favors the theory that it started from the sclerotic, a tissue which, containing but a few blood-vessels, could only furnish a scanty supply of nutritive material to the pseudoplasm. We know, however, that chondromata in general are of very slow growth. The majority of them, like the present case, are first noticed in early youth; some are congenital. The sclerotic could be recognized in this specimen with the microscope. Its outer layers appeared as usual, but the inner were infiltrated with nuclei and cells of an indifferent character, such as are found at the border of every growing neoplasm. Near by lay hyaline cartilage. There was, therefore, an immediate transition between the sclerotic and the tumor, indicating either that the cartilaginous tissue grew *from*, or grew *into* the sclerotic. The correctness of the latter view was borne out by the fact that I saw here and there stellate and angular pigment cells crowded into the sclerotic tissue. All who are familiar with intra-ocular neoplasms know how the pigment cells of the choroid move when this membrane itself is destroyed. They follow the course taken by the proliferating new elements. In spite of this, I think that the enchondroma originated in some part of the inner layers of the sclerotic, spread toward the vitreous, destroyed every tissue within the eyeball, and, after filling the globe completely, began to press and encroach upon other parts of the sclerotic, gradually expanding it by its steady increase. Thus the choroidal pigment may have

been carried with the newly-formed cells into the sclerotic when they reached and invaded the opposite side of the eyeball.

There was no indication of a special *cause* for the development of this growth. C. O. Weber, in his statistics of 237 cases of enchondroma (*Die Knochengeschwülste*, Bonn, 1856, p. 138), showed that in about half the cases where a history was given, an injury was the cause of the chondromatous growth.

R. Virchow assumes (*Morbid Tumors*, 1, p. 505) that the enchondromata of the soft parts commonly originate in the interstitial connective tissue, in which "chronic inflammatory, or at least irritative" conditions lead to the new formation of connective tissue, which is the matrix of the enchondroma. The cellular elements of the connective tissue multiply by division, the intercellular substance becomes denser and more abundant, the cells are surrounded with capsules, and the cartilage is formed (Virch., l. c., p. 506).

In the case above described there was no history of a trauma. Without any inconvenience to the patient, or irritation of the eye, the globe was noticed to enlarge, sight failing, and a white reflex shining through the pupil. This evidently was caused by newly-formed tissue in the posterior part of the eye. No other than connective and cartilaginous tissues being present in our specimen, we have no reason to disagree with the great pathologist, but may assume that the first step of the pseudoplasm was new formation of connective tissue. This we find well marked in the outer layers of the specimen, from which it runs through the tumor in

various directions, forming septa between the nodes (*d*, Fig. 1), and accumulating in larger quantity in the soft portion (*c*, Fig. 1). There is still, as we have seen, an active proliferation going on in this connective tissue (Fig. 2), resulting not only in its increase, but in its conversion into cartilage. In the outer layers of the capsule nothing is found but undulating and parallel fibres, interspersed with some nuclei; in the inner layers and the septa arising from them, the nuclei become more and more numerous, and their gradual conversion into cartilage cells can be easily observed. In the softer places of the granular portion there is a network of delicate fibres, containing nuclei and small cells (Fig. 2), but in the denser places of the granular portion the cells become larger, obtain a double outline, and one or several distinct nucleoli (Fig. 7). At some points the cells lie isolated between the nuclei and fibres of the connective tissue; at others crowded together and forming clusters walled in by a fibrous capsule (Fig. 7, *d*). Two and more cells very frequently are found in the same capsule (Fig. 7, *a*, *c*, *d*). The fibres in the basis-substance have become coarse. Thus we see fibrous cartilage at the side of proliferating connective tissue in the same microscopic field of vision (Fig. 6). In some places there are smaller and larger accumulations of cartilage cells, surrounded by connective tissue which is but little changed (Fig. 7). Such accumulations are the starting points of new nodes of the growth, which, itself, is but an agglomeration of many separated or coalescent nodes (Fig. 1).

Whereas the development of the *fibrous cartilage*

could be clearly demonstrated to consist in a *gradual conversion of the connective tissue* elements (Fig. 7), the *hyaline cartilage* showed *different modes of development.*

(1.) The *isolated clusters of cartilage cells* above mentioned (Fig. 7), formed the *nuclei of hyaline nodes*, being distinguished from the cells of the fibrous cartilage by their yellow color and lustrous appearance. The deposition of hyaline intercellular substance completed the formation of true cartilage (Fig. 7, *b*, *c*). These yellow clusters may be likened to seeds strewn into the young connective tissue, which they gradually absorb by their own growth. Some of them contain only a few cells, but have, nevertheless, thick capsules, and represent simple breed-cells (Fig. 7, *a*); others are compound clusters, consisting of a varying number of smaller clusters and isolated cells enclosed in a common large capsule (Fig. 7, *c*, *d*). The microscopic picture of these compound clusters nearly resembles the macroscopic appearance of the whole tumor (Fig. 1). The growth of the neoplasm by proliferation of cells is distinctly visible in these clusters. The matrix in which they are embedded is provided with blood-vessels, which I did not see enter into one of the clusters. In some places there were embedded into the fibrous basis-substance larger *granular bodies* (Fig. 7, *e*), which do not exactly resemble the accumulations of granular fat, the dots varying too much in size, being dark throughout, viz., without a transparent centre, and irregular in outline, unlike the little circles with white centres which compose the globules of granular fat. Here and there a

nucleus was surrounded by similar dark granules (Fig. 6, *a*), resembling a coarsely granular cell body, around which no membrane was visible.

2. *The fibrous cartilage was converted into hyaline cartilage by gradual extension of the hyaline basis substance into the fibrous matrix of the fibro-cartilage.* There were numerous yellow patches of true cartilage scattered through the fibrous cartilage. The line of transition between the two was ill-defined and often imperceptible. *The hyaline cartilage, in its growth, absorbs the fibro-cartilage as well as the connective tissue.*

3. *Agglomerated formative cells were directly converted into cartilage cells by diffuse deposition of hyaline substance.* To a large extent, especially in preparations taken from the larger nodes of the specimen, the structure of this pseudoplasm presented a dense accumulation of nuclei, or small round cells, resembling medullary tissue, for instance, glioma (Fig. 8). These small, round, formative cells (*a*, Fig. 8) were mostly of the same size, finely granular, pellucid, and colorless. Some had distinct nucleoli, others had none. Their contour was always well defined. They were pervaded, in most places, by an irregular network of, mostly, coarse fibres (Fig. 8, *b*). In the immediate vicinity of this white medullary tissue, yellow, pellucid, faintly-lustrous, small and large patches (*d*, Fig. 8) appeared, which contained the same kind of cells, embedded in hyaline intercellular substance; some of these cells, however, were shrunken. These hyaline patches extended in irregular lines (*c*, Fig. 8) into the medullary structure,

communicated with one another, broadened, and encompassed smaller and larger irregular spaces, which were densely filled with formative cells. Where the deposition of hyaline substance had only begun, its border line was commonly ill-defined, as in Fig. 8; but where it had advanced further, its boundary was sharp, and the enclosed spaces resembled breed-cells, as *b* and *c* in Fig. 3, and *c* in Fig. 4. Following these patches up a greater distance from their boundary lines, I found that the small round cells—or nuclei—enlarged, became oval, had one or several distinct nucleoli; obtained body—cell contents—and capsules around them, multiplied their nuclei, and aggregated in clusters, thus exhibiting all the features of true cartilage.

The more I studied the histology of this specimen—and I employed a good deal of time at it—the more I was convinced that the increase of the tumor was less effected by multiplication of cartilage cells in the way of cell division, but by the conversion of formative cells into cartilage cells. This process was very active, both in the fibrous and the hyaline portions of the pseudoplasm. The formative cells were either sparsely strewn into the connective tissue, or so densely crowded together that they recalled the structure of marrow, or encephaloid tumors; but here and there they were surrounded by hyaline substance, which could be easily recognized by their yellow color and lustrous appearance. Thus, as in the growth of other tumors, so in that of enchondroma, the formative cell plays the most important part.

In regard to its clinical importance, the specimen

above described must be pronounced an *innocent or benign growth.* Though in very rare instances metastatic cartilaginous tumors have been observed (Paget, Virchow), there was no indication of malignance in this case. The tumor was, moreover, completely encapsuled, even after having destroyed the eyeball long before. Its first appearance in early youth, and its slow growth, are features which it had in common with chondromatous neoplasms in general.

As an *intra-ocular tumor, it is novel and unique.* The few cartilaginous tumors to be found in ophthalmological literature were located either on the outside of the sclerotic and cornea (Von Graefe and Schweigger, *Arch.* vii. 2, p. 5), or in the orbit (Fano, *L'Union Médicale*, 1859, t. iii., p. 557, reprinted in *Demarquay's Tumeurs de l'Orbite*, p. 365, and Mackenzie, *Treatise*, 4th Edition, p. 330).

The removal of the tumor was indicated, and, if done at an earlier time, would probably not have been followed by after-hemorrhage and its fatal consequences.

I would not omit to thank Prof. Chisolm very cordially for having kindly given me the opportunity of examining so interesting a specimen, which, on account of its rarity, forms a very valuable contribution to the knowledge of ocular tumors. This being the first and only case of its kind ever observed in that locality, I took particular pains to investigate, describe, and illustrate it in such a way as to leave no uncertainty with regard to its nature.

As there are, in reality, no unique cases, I doubt not

that this observation of an intra-ocular enchondroma will soon be followed by others. H. KNAPP.

NEW YORK, Nov. 7, 1872.

EXPLANATION OF THE DRAWINGS ON PLATES A AND B.

Fig. 1. *Section through the Enchondroma.*
- *a.* Fibrous capsule.
- *b.* Hyaline nodes.
- *c.* Fibro-granular substance.
- *d.* Fibrous septa between nodes.

Fig. 2. *Proliferating connective tissue and its elementary parts.*
- *g.* Fat in granulated bodies.
- *f.* Formative cells.
- *m.* Fat in isolated globules.
- *t.* Net of connective-tissue fibres.
- *v.* Blood-vessels.
- *o.* Long fibres with double outlines.
- *s.* Spindle-shaped, *h*, stellate, and *r*, small round cells.
- *u.* Small round cells in clusters.

Fig. 3. *Hyaline cartilage.*
- *a.* Oval cell with two nuclei.
- *b.* Smaller irregular spaces containing several nuclei.
- *c.* Large irregular cavity densely filled with nuclei.
- *d.* Free nuclei.

Fig. 4. *Hyaline Matrix interspersed with free nuclei.*
- *d.* Nuclei, small, round, and pellucid.
- *b.* Nuclei, small and shrivelled.
- *c.* Nuclei, large, oval, round, club-like, dumb-bell-shaped, or irregular.

Fig. 5. *Fibrous cartilage.*
- *a.* Nuclei in a granular or finely striated matrix.
- *b.* Larger cells in a coarsely fibrous matrix.

Fig. 6. *Fibrous cartilage developing from proliferating connective tissue.*
- *a.* Nucleus surrounded by dark granules.

Fig. 7. *Smaller and larger accumulations of cartilage cells in connective tissue.*

a. Simple breed-cells.

b, *c.* Deposition of hyaline substance around the cells.

d. Large capsule.

e. Coarsely granular bodies.

Fig. 8. *Agglomerated formative cells, converting into hyaline cartilage.*

a. Small round pellucid cells, densely accumulated, as in medullary tissue.

b. Network of coarse fibres pervading the cells.

d. Yellow, pellucid, and faintly-lustrous patches, extending

c. in irregular outlines into the medullary tissue.

Erratum: The small letter *a* on the left border of Fig. 8 should be *d*.

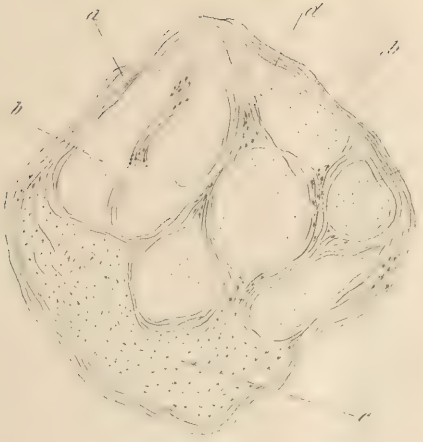


Fig. 1 a.

Fig. 2.

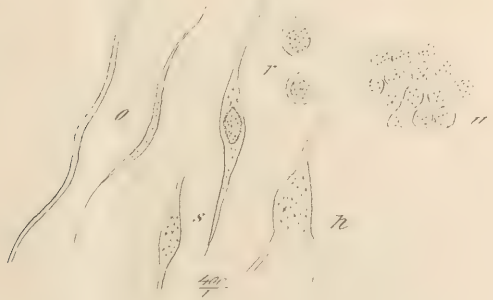


Fig. 3.

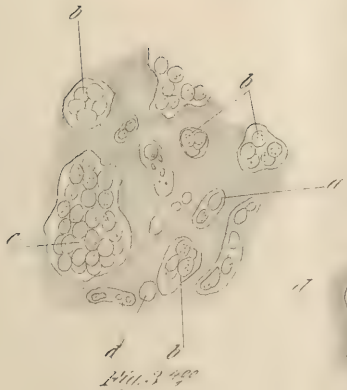


Fig. 3 a.

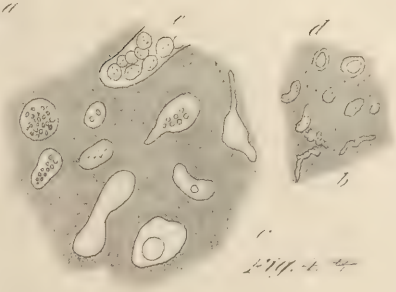


Fig. 4.

H. Knapp del.

A. Maisonneuve Lith.

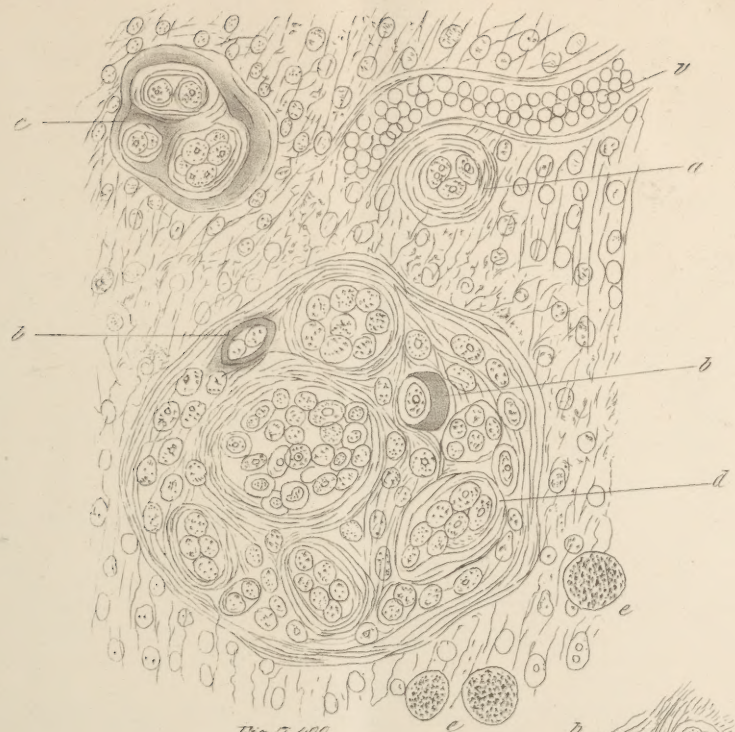


Fig. 7 400

Fig. 5 400

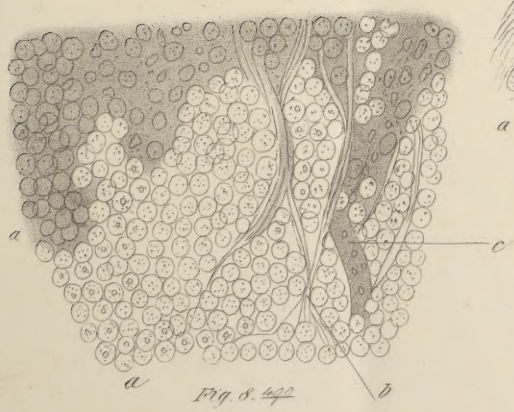


Fig. 8. 400

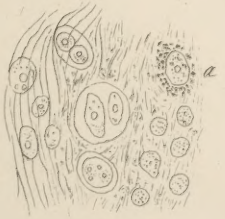


Fig. 6. 400

H. Knapp del. J. Maisonneuve Lith.